Inflammatory Breast Cancer

Editor's note: This document provides a brief description of inflammatory breast cancer. For comprehensive information on this and other types of breast cancer, please refer to our document *Breast Cancer*.

What is inflammatory breast cancer?

Inflammatory breast cancer (IBC) is rare. It is not a new type of breast cancer, but it is very important to distinguish IBC from other types of breast cancer because there are major differences in its symptoms, prognosis, and treatment.

"Inflammatory" or "inflammation" refers to changes in the body's tissues that can be caused by injury, irritation, or infection. This reaction typically involves redness, warmth, and swelling in the involved parts of the body. These symptoms are caused by increased blood flow and the buildup of white blood cells.

One type of breast cancer is called "inflammatory breast cancer" because the affected breast displays the same symptoms that may occur with inflammation, like swelling, skin redness, and breast pain. But this does not mean that IBC (or its symptoms) is caused by infection or injury. The symptoms of IBC are caused by cancer cells blocking lymph vessels in the skin.

There is some disagreement in the numbers, but IBC probably accounts for about 1% of all breast cancers diagnosed in the United States. Some experts believe that IBC may be more common, but making the initial diagnoses of IBC is often difficult. This can result in the disease not being reported as often as it should.

How is IBC different from the more common types of breast cancer?

IBC causes symptoms that are often different from those of more common breast cancers. It rarely causes a breast lump, and it may not show up on a mammogram. Because it doesn't look like a typical breast cancer, it can be harder to diagnose.

IBC tends to occur in younger women, and African-American women appear to be at higher risk of IBC than white women.

IBC also tends to grow more quickly and aggressively than the more common types of breast cancer. It is already considered to be at least stage IIIB (*locally advanced*) when it is first diagnosed, and may be stage IV (*metastastic*) if it has spread to distant parts of the body. Because IBC is always diagnosed at a high stage, it is often harder to treat successfully than other types of breast cancer.

What are the signs and symptoms of inflammatory breast cancer?

Common signs and symptoms of inflammatory breast cancer (IBC) can include:

- Breast swelling, which is usually sudden with one breast much larger than the other
- Itching (of the breast)
- A pink, red, or dark colored area, sometimes with a texture like the skin of an orange (on the breast)
- Ridges and thickened areas of the skin (of the breast)
- Breast feeling warm to the touch
- Nipple retraction
- Breast pain

Tenderness, redness, warmth, and itching are also common symptoms of a breast infection or inflammation (such as *mastitis*). Because these conditions are much more common than IBC, a doctor may at first suspect them as the cause. This may delay the true diagnosis. A long delay can give the cancer a chance to spread. By the time cancer is diagnosed, lymph nodes may be enlarged under the arm or above the clavicle (collar bone).

If you have any of these symptoms, it does not mean that you have IBC, but you should see your doctor without delay.

Can inflammatory breast cancer be detected by mammogram or breast examination?

Because of the way inflammatory breast cancer (IBC) grows and spreads, a distinct lump may not be noticeable during a clinical breast exam, breast self-exam, or even on a mammogram. However, signs of IBC can be seen on the surface of the skin, and skin thickening often shows up on a mammogram and can be seen during a clinical breast exam or breast self-exam.

Symptoms of IBC can develop very quickly, so women should pay attention to how the skin on their breasts looks and tell their doctors about any changes in skin texture or breast appearance.

In women who are pregnant or breastfeeding, breast redness and swelling is more often caused by an infection than by IBC, so doctors may first try treatments like antibiotics for a short time. The possible diagnoses of IBC should be considered when a woman who is not pregnant or breast feeding comes in with these symptoms. Breast infection is very rare in women after menopause. When infection occurs it is usually associated with fever or other signs of infection. If treatment with antibiotics is started you will need to keep your doctor informed if this treatment doesn't help, especially if your symptoms worsen or the area affected gets larger. Ask for a referral to a specialist or seek a second opinion if you are concerned.

Following American Cancer Society guidelines for early detection of breast cancer can improve a woman's odds of finding breast cancer (especially the usual forms of breast cancer, but also IBC) as early as possible, when it can be treated most successfully

- Women age 40 and older should have a screening mammogram every year and should continue to do so for as long as they are in good health.
- Women in their 20s and 30s should have a clinical breast exam (CBE) as part of a periodic (regular) health exam by a health professional, preferably every 3 years. After age 40, women should have a breast exam by a health professional every year.
- Breast self-exam (BSE) is an option for women starting in their 20s. Women should be told about the benefits and limitations of BSE. Women should report any breast changes to their health professional right away.
- Women at high risk (greater than 20% lifetime risk) should get an MRI with their mammogram every year. Women at moderately increased risk (15% to 20% lifetime risk) should talk with their doctors about the benefits and limitations of adding MRI screening to their yearly mammogram. Yearly MRI screening is not recommended for women whose lifetime risk of breast cancer is less than 15%.

(For more information on the American Cancer Society guidelines, see our document, *Breast Cancer: Early Detection*.)

Because IBC grows and spreads so fast, screening is not as helpful for finding this disease early.

How is inflammatory breast cancer diagnosed?

Imaging studies

If inflammatory breast cancer (IBC) is suspected, a mammogram may be the first test ordered. Sometimes the tenderness and pain of IBC may make it hard to do a good

mammogram. Often a breast ultrasound is ordered as well. The mammogram may show thickened skin, often without a visible mass (tumor). It may also show that the affected breast is larger than the other as well as increased breast density. The ultrasound is often able to show that lymph nodes under the arm are enlarged and may find breast masses (tumors) if they are present. Ultrasound can also be useful in guiding a needle for a biopsy procedure.

MRI (magnetic resonance imaging) is the most sensitive imaging test for IBC. It can find any breast mass that looks like it could be invasive cancer. More importantly, MRI allows the skin changes that are typical of IBC to be measured precisely.

Another test that may be useful is called a PET (positron emission tomography) scan. This test is often combined with a CT (computed tomography) scan. It can be useful in finding areas of cancer spread to lymph nodes and distant sites. PET/CT is one of the best ways to find IBC that has spread to the nearby and distant lymph nodes, which are among the most common sites of IBC spread.

It is important that every woman with a diagnosis of IBC has a photo of the breast taken before starting treatment. It is best if the photo is taken at the same time as the imaging studies. This approach can help the doctor learn how much cancer is present and can be used as a comparison to see if the cancer has responded to treatment.

More information about these tests can be found in our document, *Breast Cancer*.

Biopsy

Like any other type of breast cancer, the diagnosis of IBC is made by a biopsy -- removing a sample of the breast tissue and looking at it under the microscope. Other tests may show findings that are "suspicious for" IBC, but only a biopsy can tell for sure that cancer is present. Breast biopsies can be done in many ways. Samples of breast tissue can be removed using fine needle aspiration, large core biopsy, vacuum assisted biopsy, or open (excisional or incisional) biopsies -- depending on where the affected area is, what it looks like, and who finds it. A biopsy in IBC often involves a needle biopsy under ultrasound guidance. IBC can also be diagnosed with a skin biopsy.

How aggressive is inflammatory breast cancer?

Inflammatory breast cancer (IBC) is said to be an aggressive cancer because it grows quickly and it is more likely to have spread to nearby lymph nodes at the time it is found than other types of breast cancer. The prognosis (outlook) is generally not as good as it is for other types of breast cancer.

Doctors often use *5-year survival rates* as a way to discuss prognosis in people with cancer. The 5-year survival rate refers to the percentage of patients who live at least 5 years after being diagnosed with cancer. (Many of these patients live much longer than 5 years.) Five-year *relative survival rates*, such as the numbers below, compare the number

of people who are still alive 5 years after their cancer was found to the survival of others the same age who don't have cancer. They are considered to be a more accurate way to describe the outlook for patients with a particular type of cancer.

According to data from the National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) database, for women who were diagnosed with inflammatory breast cancer between 1988 and 2001, the 5-year relative survival rate was about 40%. This compares with about 87% for all breast cancers combined.

These numbers are among the most current available, but they were based on patients treated at least several years ago. Improvements in treatment since then mean that the survival rates for women now being diagnosed with these cancers may be higher.

Survival statistics can sometimes be useful as a general guide, but they may not accurately represent any one person's prognosis. A number of other factors, including other tumor characteristics and a person's age and general health, can also affect outlook. Your doctor is likely to be a good source as to whether these numbers may apply to you, as he or she is familiar with the aspects of your particular situation.

How is inflammatory breast cancer treated?

The usual treatment for inflammatory breast cancer (IBC) starts with chemotherapy (chemo). Chemo is the use of drugs for treating cancer. The drugs can be swallowed in pill form, or they can be injected by needle into a vein or muscle. Because the drugs enter the bloodstream and circulate throughout the body to reach and destroy cancer cells wherever they are, chemotherapy is considered systemic therapy. Systemic treatments can destroy any cancer cells that break off from the main tumor and travel in the bloodstream to lymph nodes or distant organs.

Using chemotherapy before surgery is called *neoadjuvant* chemotherapy. Anthracyclines (doxorubicin or epirubicin) and taxanes (paclitaxel or docetaxel) are the most effective chemotherapy drugs for IBC, and most women with IBC receive a combination of at least two different drugs. Some combinations that may be used include:

- CAF (FAC): cyclophosphamide, doxorubicin (Adriamycin®), and 5-fluorouracil
- AC: doxorubicin (Adriamycin) and cyclophosphamide
- EC: epirubicin (Ellence®) and cyclophosphamide
- TAC: docetaxel (Taxotere®), doxorubicin (Adriamycin), and cyclophosphamide
- AC → T: doxorubicin (Adriamycin) and cyclophosphamide followed by paclitaxel (Taxol®) or docetaxel (Taxotere).
- CEF (FEC): cyclophosphamide, epirubicin, and 5-fluorouracil (this may be followed by docetaxel)

If the cancer is HER2-positive (the cancer cells have too much of a protein called HER2), the targeted therapy drug called trastuzumab (Herceptin®) is given. This drug can lead to

heart problems when given with an anthracycline, so often it is given with the drugs docetaxel (Taxotere) and carboplatin in a combination known as TCH.

Aggressive chemotherapy is often followed by treatment aimed at the breast, also known as local regional treatment. The 2 major types of local treatment are surgery and radiation. Surgery is used if the cancer has not spread too far to be removed completely. Sometimes chemo given before surgery will cause the cancer to shrink enough so that it can be removed with surgery. The usual operation is a modified radical mastectomy, where the entire breast and the lymph nodes under the arm are removed. Because IBC involves so much of the breast and skin, a lumpectomy or skin-sparing mastectomy is not a treatment option. If, after chemotherapy and surgery, no cancer is found in the breast or in the lymph nodes, the patient is far less likely to have the cancer recur (come back).

Breast radiation is used in most cases, whether or not surgery is done, to further slow the disease. Radiotherapy is usually given once a day for 6 weeks, in some cases a more intense treatment (twice a day) can be used in this disease. Radiotherapy may be followed by additional systemic treatment. This is known as *adjuvant therapy* and can include chemotherapy, hormonal therapy (tamoxifen or an aromatase inhibitor, if the cancer cells contain estrogen receptors), and/or trastuzumab (if the cancer is HER2-positive).

For more information on breast cancer treatment, see the "How Is Breast Cancer Treated?" section of our document, *Breast Cancer*.

What's new in inflammatory breast cancer research?

Because inflammatory breast cancer (IBC) is so rare, it makes it harder for researchers to find women to study and find the best treatments for it. But there have been some recent advances in understanding and treating IBC.

Studies have shown that over the past couple of decades, IBC has become more common, while other forms of locally advanced breast cancer have become less common. Researchers are still not sure why this has happened.

Studies comparing DNA and other molecules from IBC with that of the usual types of breast cancer have shown some important differences. Scientists believe that some of these differences are responsible for the unique and aggressive way that IBC spreads and grows. They are hopeful that understanding these differences will lead to more effective treatments that target molecules specific to IBC.

Clinical studies during the past decade have shown doctors how to modify the usual breast cancer treatments (chemotherapy, radiation, hormonal therapy, and surgery) so that they are best suited for women with IBC. For example, studies have shown the value of using chemotherapy that is more intense than the usual regimens for breast cancer, and the importance of using chemotherapy as the first treatment, before surgery or radiation.

A study has shown that lapatinib (Tykerb) can be a useful treatment for IBC in women whose disease has stopped responding to regular chemo plus trastuzumab. In this study

the lapatanib was given alone, without other chemo drugs. It caused the tumors to shrink in many of the women treated. Further studies using this drug to treat IBC are going on now.

Where can I find more information about inflammatory breast cancer?

Inflammatory Breast Cancer Research Foundation
Telephone: 251-866-0907
Web site: www.ibcresearch.org

For more information about breast cancer, please see our documents, *Breast Cancer* and *Breast Cancer: Early Detection*.

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